

Pediatric Rounds

Participants

ELMER E. SPECHT, MD
ERNEST K. GOODNER, MD
EMIL A. TANAGHO, MD, M Ch
BARBARA PRINCE, MSW
BYRON C. PEVEHOUSE, MD
PETER COHEN, MD

*Presented by the Spina Bifida Group
of the University of California,
San Francisco*

Refer to: Specht EE, Goodner EK, Tanagho EA, Prince B, Pevehouse BC, Cohen P: Myelomeningocele—A symposium on orthopedic, ophthalmologic, urologic, psychological and social, neurosurgical and general considerations (Pediatric Rounds). West J Med 121:281-304, Oct 1974

Myelomeningocele

A Symposium on Orthopedic, Ophthalmologic, Urologic, Psychological and Social, Neurosurgical and General Considerations

Refer to: Specht EE: Myelomeningocele—Part I: Orthopedic management in children, *In* Myelomeningocele—A symposium. West J Med 121:281-291, Oct 1974

MYELOMENINGOCELE—PART I

Orthopedic Management in Children

ELMER E. SPECHT, MD, *San Francisco*

IF AN INFANT with myelomeningocele survives, the goals of orthopedic management are (1) a straight spine and straight legs and feet, (2) mobile hips, (3) a pull-to or swing-through gait and (4) maximum possible self-sufficiency, including whatever can be accomplished by vocational training at an appropriate age. Helping the patient to attain these goals depends on informed evaluation of the expected functional capacity in relation to a given neurological deficit, and assessment of the likeli-

hood of fixed deformities related to muscle imbalance and positional contractures. Familiarity with techniques for correction of deformities and for habilitation, with specific reference to walking—if that is a realistic goal—are also essential.

One of the most important responsibilities of the orthopedic surgeon evaluating these children is to determine the pattern of remaining neurological function and the presence of functioning or spastic muscle groups, as well as the status of their antagonists, and to anticipate subsequent deformity (Table 1). It does not necessarily follow from the fact that an infant has been born with myelomeningocele that there will be handicapping neurological impairment. In a study published in 1961, Doran and Guthkelch¹ reported 307 cases followed for a minimal period of two and a half years. They found that of 95 survivors after treatment of myelomeningocele with and without hydrocephalus, 66 showed either no paralysis or no more than minor weaknesses in the lower extremities, even though sphincter paralysis was frequently present. Comparable findings were more recently reported by Piggot,² in whose series 32 of 108 babies with myelomeningocele (but without spinal fluid leak and without hydrocephalus at birth) showed no detectable muscle weakness at birth.

Part I of an article in six parts.

From the Department of Orthopaedic Surgery, University of California, San Francisco.

Reprint requests to: E. E. Specht, MD, Department of Orthopaedic Surgery, University of California, San Francisco, San Francisco, CA 94143.

MYELOMENINGOCELE

TABLE 1.—Correlation of Neurological Level, Remaining Muscular Function, and Deformity or Disability Most Commonly Encountered in Myelomeningocele

Level Intact	Principal Motor Groups Remaining Functional in Legs	Principal Motor Groups Paralyzed or Weak in Legs	Deformities and/or Disabilities of Spine and Legs Due to Muscle Strength Imbalance (Positional Deformities May Be Superimposed)
T ₁₂	None	Complete lower-extremity paralysis	Spinal scoliosis or lordosis Coxa valga, occasional hip subluxation
L ₁ -L ₂	Hip flexors fair to good Hip adductors poor to fair	Hip extensors paralyzed Hip abductors paralyzed Knee and foot flail	Spinal scoliosis or lordosis Flexion-adduction contracture of hip, with subluxation frequently, dislocation occasionally
L ₃ -L ₄	Hip flexors normal Hip adductors good to normal Quadriceps fair to good Tibialis anterior trace to fair	Hip extensors paralyzed Hip abductors paralyzed Knee flexors paralyzed Triceps surae paralyzed Foot evertors paralyzed Foot invertors paralyzed	Lumbar lordosis or scoliosis Flexion-adduction-external rotation contracture of hip early; hip dislocation at birth or early Extension contracture of knee occasionally Severe, progressive calcaneus of foot
L ₅	Hip flexors and adductors normal Quadriceps normal Medial hamstrings fair to good Foot dorsiflexors and invertors normal	Hip extensors trace to poor Hip abductors poor to fair Triceps surae paralyzed Foot evertors fair	Lordosis, scoliosis Flexion-adduction contracture of hip; coxa valga, subluxation or dislocation common Knee flexion-internal rotation contracture Severe progressive calcaneovarus of foot
S ₁ -S ₂	Hip nearly normal except extensors Knee good except lateral hamstrings Foot and ankle extrinsics good to normal except toe flexors	Hip extensors fair to good Biceps femoris fair Foot intrinsics paralyzed Toe flexors fair	Lordosis, scoliosis Occasional subluxation of hip Planovalgus foot, claw toes

In the treatment of deformity due to myelomeningocele, it is vital to recognize that the deformity caused by dynamic, occasionally spastic,³ muscle imbalance differs from the relatively static situation one often encounters in deformities caused by old poliomyelitis. Deformity may progress slowly because a minimal imbalance of forces may be acting over a prolonged period of time. The concept of a child who is a *growing paraplegic* is useful in understanding this tendency to slow progression of deformity. Serial evaluations throughout childhood are essential.

Deformities of the Foot and Ankle

Orthopedic management of the foot (as well as of the spine, hips and legs) in myelodysplasia must always be directed toward the ultimate goals of the educability and employability of the patient. A mobile, plantigrade, balanced, shoeable foot is essential, and one of the responsibilities of the orthopedist, as elsewhere in the management of

this disorder, is to determine the level or pattern of remaining neurological function and to predict the type of deformity which this pattern can be expected to produce. Thus, with a functional fourth lumbar level, the tibialis anterior muscle may be innervated, but its action may be unbalanced by functioning plantar flexors and evertors. A varus force may be produced; but more important, a progressive and severe calcaneus deformity may develop, which can be minimized by early transfer of the tibialis anterior tendon to the heel. With a functional fifth lumbar level, a calcaneus deformity will almost certainly develop if tendon transfer is not done by the time the patient is one to two years old, and at present no satisfactory means of managing a fully developed calcaneus deformity exists. At the first and second sacral levels, ankle plantar flexors and foot evertors will be functioning, so that clawing of the toes and a planovalgus foot are more common deformities. At the third sacral level, intrinsic muscular

imbalance obtains and a cavus foot is most likely. The equinus portion of the deformity that is commonly present is a positional rather than a dynamic problem and is thus less apt to recur after tenotomy. All dynamic deformities, however, must be brought into balance by neurectomies, tendon transfers or tendon releases; otherwise recurrence is a certainty.

The two deformities of the foot in myelodysplasia which most commonly require correction are equinovarus and equinocavovarus.⁴⁻⁶ Opinion on the management of these deformities varies. Sharrard⁶⁻⁹ feels that splinting, manipulation and serial plaster casting for these insensitive feet result in disaster. Hayes, Gross, and Dow,⁵ Tzimas¹⁰ and Walker,¹¹ on the other hand, believe that such measures are useful. However, all of these authors warn against the injudicious use of plaster on feet in which sensation is lacking, and all resort to the use of surgical procedures if necessary in order to obtain correction. Sharrard and Grosfield⁶ have reported the results of their management of 241 feet with various deformities; they found soft-tissue releases and tendon transfers to be most effective. For equinovarus deformity they advised medial and posterior releases; their rate of success in this group was 22 percent. Usually, the tendo achillis, both long toe flexors, both tibial tendons and the medial ligaments of both the ankle and the subtalar joints will require division. An unopposed tibialis anterior tendon should be transferred to the lateral side of the foot if no tendency to calcaneus deformity is present; if it is, the tendon should be transferred posteriorly. For simple equinus contracture, subcutaneous tendo Achillis tenotomy and toe flexor tenotomy will often suffice.

Talectomy may also be of value in the management of rigid feet in which equinovarus deformity cannot be controlled and the potential for walking is limited. Menelaus¹² reported on his use of this procedure in 14 feet with equinovarus deformity due to spina bifida. He believes that best results can be obtained if talectomy is done between the ages of one and five years, but that triple arthrodesis is preferable for the older child or adult.

Planovalgus and equovalgus feet are also commonly seen in myelodysplasia, and convex pes valgus (vertical talus) has been described.¹³ The Grice subtalar arthrodesis is useful in the correction of planovalgus deformity. Paralytic vertical talus is a difficult deformity to manage. Dorsolateral release with open reduction is indicated,

along with such tendon transfers as are necessary to bring the musculature of the foot into balance.

The fully developed calcaneus and calcaneovarus deformities which the clinician occasionally encounters are among the most difficult to correct. In cases in which an unbalanced overpull of the peroneus tertius, the tibialis anterior, or both, cause an elevation of the arch, both tendons should be transferred through the interosseous membrane to the heel at the midline of the foot. These tendons are usually not sufficiently strong to act as plantar flexors or to cause equinus deformity, but in the event that equinus ensues, this deformity is more easily held by a dropfoot brace, whereas a calcaneus deformity cannot be held by any brace. These transfers should be done when the child is about a year old.

In order to salvage some feet, triple arthrodesis may be necessary, but this procedure should not be undertaken lightly. Nonunion after attempted triple arthrodesis has occurred in nearly one-third of cases in some series.⁵ It should also be borne in mind that in the past, at least, osteomyelitis in insensitive feet has often required amputation in order to control the infection. Pantalar arthrodesis should be avoided; unwanted ankle motion can be controlled by the placement of appropriate anterior and posterior stops on short leg braces. Neuropathic arthropathy may occur after attempted arthrodesis of joints, especially the ankle; furthermore, even if fusion is accomplished, there is no virtue in a completely rigid ankle and hindfoot.

Finally, it must be stressed that nonplantigrade feet, particularly those that have been immobilized in plaster casts, but also those that have been supported by braces for prolonged periods of time, are vulnerable to trophic ulceration. Casts, if they are used at all, must be well padded and bivalved; removable splints are preferable. Skin should be inspected for rubor at pressure points at least twice daily. It is better to maintain a surgically induced correction by passive movement of the involved joint than by the use of plaster. It is also better to obtain a correction by passive manipulation and to maintain it by plaster splints, if necessary, than to attempt plaster correction of deformity as one might in congenital clubfoot with normal skin sensation.

Deformities of the Knee

Although Carr⁴ states that the most common knee deformity in his series was knee flexion contracture, which he attributed to strong hamstrings

and calf muscles opposed by a weak quadriceps, it is generally agreed that hyperextension deformity occurs more frequently, especially when the third lumbar segment is functioning; the result is a nearly normal quadriceps with weak knee flexors. Sharrard⁹ advocates early elongation of the quadriceps to allow at least 45 degrees of knee flexion in severe cases. Hayes and associates, on the other hand, feel that serial plaster casting in infancy will allow sufficient correction that a long leg brace will permit ambulation later. They recommend transfer of the hamstring tendons to the femoral condyles in those instances of knee flexion deformity due to myelodysplasia with spasticity, as in cord traction syndromes. Derotational osteotomy of the tibia is occasionally indicated. Supracondylar femoral osteotomy for persistent contracture in patients over the age of eight may also be useful, and of more permanent benefit, than simple posterior knee release for positional flexion contracture.

Paralytic Dislocation of the Hip

The problem of paralytic dislocation of the hip has been studied in considerable depth by Sharrard,¹⁴ who found that hip dislocation did not occur if there was complete paralysis of all hip musculature. If the first, or first and second, lumbar segments were functioning, moderate or severe subluxation, in association with progressive flexion-adduction deformity and coxa valga, developed in the first year of life in 58 of 70 hips. An additional eight in this group were dislocated.

The most striking incidence of dislocation, however, occurred in the group in which the upper three or four lumbar roots were intact, with strong hip flexors and adductors but complete paralysis of the extensors and abductors. In this group, dislocation was either present at birth or developed in the first year of life in 72 of 85 limbs, and the remaining 13 were subluxed, in spite of normal development of the acetabulum.

In the group in which all five lumbar segments were functioning normally, with good hip musculature except for weak abduction and paralyzed extension, the picture was similar to that in which the first and second segments were functioning; a slowly progressive flexion-adduction contracture developed, with subluxation of 12, and dislocation of 7, limbs in the group of 27 with this pattern. If hip extension only was weak, no deformity was seen at birth except for mild fixed flexion in some infants, who did not, however, have dislo-

cated hips. No subluxation or dislocation was seen when innervation of the limb was intact.

Sharrard's proposed remedy for this problem was posterolateral transplantation of the iliopsoas. In 1964 he reported¹⁴ that he had done 150 such procedures on children between the ages of four months and 17 years, the mean age being three years. His initial report was quite enthusiastic, and numerous "Sharrard procedures" have been done subsequently.¹⁵ In a recent long-term follow-up study,¹⁶ however, it became evident that this procedure probably has very limited applications. Indeed, one prominent children's orthopedist has recently termed it "the necrotizing iliopsoas transfer." Carroll and Sharrard's recent findings were that instability of the hip persisted in 40 percent of cases at review five to ten years postoperatively, and that in the absence of gluteal muscles the transplanted muscle was unlikely to provide abduction or extension against gravity (in other words, it had poor-grade strength). They state that the most consistent reason for late instability was failure to obtain a concentric reduction at the time of operation. They feel it important to restore muscle balance and, in addition to carrying out indicated procedures, to correct dysplasia or maldirection of either the acetabulum or the femur, as well as to correct coxa valga. In this context it should be stated that Salter¹⁷ does not feel that his technique of innominate osteotomy is indicated in the paralytic dislocation of myelomeningocele. Carroll and Sharrard's conclusion was that with high paraplegia, neonatal dislocation, and acetabular dysplasia, the 25-percent risk of avascular necrosis did not justify more than such soft-tissue releases as are necessary to permit bracing. They continue to feel that, with lower lumbar levels of paralysis and a better prognosis for ambulation, the procedure is justified. Similar findings have been reported by Rueda and Carroll¹⁸ in 21 children with 30 iliopsoas transfers, 15 by the Sharrard and 15 by the Mustard technique. In this series only 10 of the 30 hips operated on remained stable at review one to eight years after transfer, and active power of abduction was poor. A surprising finding was that the power of hip flexion remained strong in 22 hips in spite of transfer of the major hip flexor.

Cruess and Turner¹⁹ in 1970 reported their results with the use of Mustard's iliopsoas transfer through an anterior window cut in the ilium. Their indications were (1) severe abductor lurch in the presence of a strong hip flexor, (2) flexion con-

tracture of the hip due to muscle imbalance and (3) muscle imbalance leading to progressive valgus of the femoral neck. It must be noted that they list two contraindications to the procedure: (1) absence of other active hip flexors and (2) subluxation or dislocation of the hip. In either of these cases the transfer should be done only as an adjunct to some procedure that corrects the posterior acetabular defect. Eight of 13 somewhat older patients, 7 to 15 years of age, were benefited functionally, according to this report, in spite of the fact that all transferred muscles lost some force. Essentially similar results were reported by Freehafer, Vessely and Mack²⁰ in a larger series in which they used a modification of the Mustard technique. They stated that all but seven of 52 hips improved, but it was their impression that the success of the transfer was more related to removal of a deforming force than to the strength of the transferred tendon. Broome and Basmajian²¹ have shown that electrical activity of the transferred muscle can be lost; three of seven transfers resulted in greatly reduced activity.

Hayes, Gross and Dow⁵ have had a similar experience in treatment of paralytic dislocation of the hip. They concur with Sharrard that varus osteotomy as suggested by Jones²² and the rotation-adduction osteotomy advocated by Somerville²³ are not adequate in the face of muscular imbalance. Donaldson²⁴ and Tzimas¹⁰ have also advocated iliopsoas transfer and have used it in conjunction with Salter's innominate osteotomy.

Hogshead and Ponseti²⁵ have approached the same problem in a different manner. It is their opinion that dislocation of the hip is related to contracture of the iliotibial band rather than to muscle imbalance. Proceeding on this theory, they have transferred the distal end of the iliotibial band subfascially across the buttock and sutured it to the lumbodorsal fascia just lateral to the spine at the level of the third or fourth lumbar vertebra. The proximal end is left anchored to the greater trochanter, and an anterior hip release is done at the same time if there is any flexion contracture of the hip. In addition they incise the lumbodorsal fascia if there is fixed lumbar lordosis. They had done this in nine cases of myelomeningocele when they reported in 1964. Their patients were between three and a half and ten and a half years of age at the time of surgery, and they stated that they obtained an average of 79-percent relief of the flexion deformity. Of the 13 hips which were

dislocated or subluxed preoperatively, 11 were improved and only one remained dislocated. They have found that the procedure results in a dynamic fasciodesis, but that few of the patients were able to discard canes or crutches after the operation. They claim, however, that better stance and easier gait result. No recent follow-up on this procedure has been forthcoming.

Others have suggested that transfer of the adductors to the ischial tuberosity in addition to transfer of the iliopsoas may be necessary to bring the hip into stable muscle balance. All authors agree that whatever is done, mobility of the hip must be maintained at all costs, and it would appear that dislocated mobile hips are preferable to reduced immobile ones. It may even be that a much more conservative approach will prove useful; in a preliminary report, McKibbin²⁶ has expressed the opinion that abduction-internal rotation splinting from an early age may obviate the need for surgery in some cases. It should always be recalled that as adults, very few if any patients with bilateral paralysis below the second lumbar level will be functional walkers, and that if surgery of this magnitude is contemplated, it should be so timed that school progress is not interfered with. Children with one or both hips dislocated can still ambulate well, and although stability is highly desirable, a minimum of 90 degrees of flexion is essential.

Deformities of the Spine and Pelvis

Spinal and pelvic deformity in childhood and adolescent paraplegia have been extensively studied by Kilfoyle, Foley and Norton.²⁷ In 1965 they reported their experiences with 107 children and adolescents who had been admitted for either congenital or acquired paraplegia. Of this group, 73 had congenital paraplegia, and of the entire group all but 10 had significant spinal deformities. The authors stated that with deforming forces at work from birth, every possible deformity in the spine and pelvis was encountered. These ranged from kyphoscoliosis so severe that only the prone position could be assumed to minimal excessive lumbar lordosis. Forty-seven of the deformities were lordosis, scoliosis or kyphosis alone, and the remainder were combinations of the three, with one being the major deformity.

In this series, lordosis was the predominant or major deformity, and it occurred in 47 of the 97 patients with spinal deformity. In a total of 80 there was some lordotic deformity of greater or

lesser degree, either alone or in association with other deformities. The authors found that there was a distinct correlation between the level of the lordosis and the state of balance. In 19 patients with lower-level lordosis and paralysis at or below the third lumbar segment, none were completely off-balance and only five of the group were found to be partly balanced. All of these five had gluteus maximus paralysis and four had either dislocation or subluxation of the hip. In the group of patients with spinal cord involvement from the third lumbar segment down to the sacrum, lordosis, hip flexion contracture and dislocation are the preponderant deformities.

In this series there were 36 patients who had scoliosis as a predominant deformity. Of these, two were well balanced, 27 were partially balanced, and seven were completely off-balance (unable to sit or stand without support). The authors noted that in the partially balanced patients the ability to walk was affected primarily by the lordosis, whereas the scoliosis had a direct effect on the ability of the patient to sit easily and use both hands while sitting. Of the seven patients who were completely off-balance, all but one were so deformed and rigid that correction was impossible. These authors believe that this state of affairs is preventable. An attempt to relate the level of paraplegia to the incidence of scoliosis showed that there was no particular correlation and that the frequency of this deformity was about equal at all levels of paralysis. In nine patients the curve could be attributed to anomalous vertebral development with angulation. In others a chronically assumed position appeared to be a significant causative factor. In our opinion, pelvic obliquity is especially to be watched as a predisposing influence. Raycroft and Curtis,²⁸ in the series at the Newtonington Children's Hospital, found that, in those patients who had no curve at birth, more than half of those who subsequently developed scoliosis did so before age five, and almost all by age ten.

Fourteen of the 97 patients in the series of Kilfoyle and associates²⁷ had kyphosis and, of these, seven had congenital paraplegia. Progression of the deformity during the years of growth is common. Paraspinal muscles which become displaced anteriorly to the center of rotation of the vertebral body at the apex of the kyphosis begin to act as spine flexors, and the superincumbent weight of the upper trunk and head become more effective forces as the lever arm of the cephalad

portion of the spine becomes longer with growth. Standing balance may become seriously impaired or lost. Severe kyphosis may result from failure of formation of an ossification center for a vertebral body. In this case the bodies above and below converge in the space which should be occupied by the anterior half of the missing body, and this kyphosis increases with growth and becomes a hairpin curve. If allowed to progress to this extent, such a dorsal hemivertebra inevitably results in off-balance kyphosis and inability to stand, even with external support.

Rope Spine

A completely flaccid, or rope-like, spine may be seen in some instances of high-thoracic paraplegia. The spine collapses or telescopes when the patient is sitting or attempting to stand, and no external support devices are adequate. This would appear to be a clear-cut indication for spinal fusion.

Treatment of Spinal Deformity

The most commonly encountered problem is the combination of hip-flexion contracture and increased lumbar lordosis. Anterior hip release of the Soutter type, with release of all contracted structures including the iliopsoas, is indicated if there is any compromise in balance, that is, if the patient cannot stand momentarily in braces without the support of the hands on crutches. We favor a two-team approach to both hips at the same time, followed by a double spica cast for no more than three weeks to allow healing with the hips extended, and then vigorous gait training. During the period in the cast, the patient should be stood upright as much as possible—a minimum of once daily.

Treatment of scoliosis is indicated if there is any reason to suspect that a curve will progress to the point that sitting balance will be impaired. There are no hard data on the method of choice for management except that the problem of insensitive skin makes the Milwaukee brace an inappropriate solution. Most authors have favored Harrington rod instrumentation in recent years. In the future, the Dwyer anterior interbody technique may prove to be the best, in experienced hands. However, its use at the present should be limited to those centers which are studying its effectiveness. Fusion to the sacrum with the use of a sacral bar appears to be indicated if there is pelvic obliquity. Casts should not be used post-

operatively. Fusion and infection rates do not appear to be substantially different from those encountered after surgical procedures on nonparalytic curves.

Treatment of kyphosis in association with spina bifida remains an unsolved problem. Sharrard in 1968²⁹ reported good results in six newborn infants in whom he had done closing-wedge spinal osteotomies for severe kyphotic deformities of the lumbar spine in order to effect skin closure of the neural defect without tension. In a more recent report,³⁰ Sharrard and Drennan state that they have done seven more, with only one death in the intraoperative period. In two patients, the procedure failed to achieve bony union and reoperation was required. Skin problems improved in all, but there was a tendency to slow recurrence of the deformity, at a diminished rate of progression, which they attribute to the perverted flexor action of the anteriorly displaced spine extensors. James and Lassman,³¹ on the other hand, have held that gross lumbar kyphosis is a contraindication to any early operative intervention.

Eyring and Wanken³² reported in 1972 on spinal osteotomy for kyphosis in 15 children ranging in age from one day to nine and a half years. Their goals were (1) to eliminate total skin ulceration, (2) to permit supine lying and alignment of legs, (3) to permit brace wearing and ambulation and (4) to improve appearance. No patient's condition was completely corrected, although all were improved; there were no deaths and no patient lost ambulatory ability. They stated that the average blood loss during the first ten procedures was 120 percent of total blood volume, and conceded that it is "a reasonably hazardous procedure." MacEwen,³³ on the other hand, has said that he knows of several deaths caused by attempted late spinal osteotomy, and he considers it simply "hazardous." This opinion would seem to be borne out by a recent paper by Eckstein and Vora,³⁴ in which they report 16 cases, in patients ranging in age from newborn to eight years, with five deaths. In spite of the high mortality, they concluded that spinal osteotomy is justifiable and reasonable in cases of severe kyphosis complicated by skin problems, inability to fit braces, reduction of abdominal area such that urinary diversion cannot be done, and reduction of abdominal volume to the extent that elevation of the diaphragm with compromise of thoracic volume are significant. Sharrard and Drennan³⁰ also recently reported the results of spinal osteotomy in 18 additional older children

(average age seven years), for similar indications, without intraoperative mortality. Union of the osteotomic site occurred in all cases, and the average correction was 33 degrees; the average loss of correction two years after operation was 11 degrees. Skin problems were said to have improved in all but two of 14 children; two who were previously unable to sit in a wheelchair were able to do so at follow-up.

It would appear to us that this procedure is justifiable only in the newborn period in cases of severe deformity, as an aid to skin closure, and in very rare circumstances in older children.

Complications of Orthopedic Management

Failure of attempted arthrodesis is a common occurrence in the management of patients with myelomeningocele. Osteomyelitis as a complication of bone operations on the feet occurred in roughly 6.6 percent of the patients in the series of Hayes and associates.⁵ Kilfoyle and associates,²⁷ on the other hand, reported no incidence of infection in 19 spinal stabilization procedures.

Impaired sensation with resulting trophic ulceration is a common complication in the management of these patients and is commented on by numerous observers. Carr,⁴ in an earlier series, stated that the incidence over a period of some years was 50 percent in patients who had sensory loss. The common sites were the posterolateral aspect of the heel, the outer side of the foot, the dorsum of the toes, and the buttocks. In older series, 50 percent of patients in whom such ulcers developed on the feet ultimately had to have amputation. Hayes, Gross and Dow,⁵ in reviewing their 100 cases, found that amputation below the knee or more distally had occurred in 13 cases. The causes included infection (most commonly), severe deformity, fractures, and Charcot joint. In 24 of their patients, trophic ulcers significant enough to require operative treatment occurred. Bluestone and Deaver³⁵ in their experiences with 45 children reported decubitus ulcers in 19 patients. In six cases the lesions were regarded as serious. Numerous observers stress the need for frequent examination of the skin for pressure areas.

Neuropathic arthropathy was found by Hayes and associates⁵ in eight of 100 patients—in the ankle joint in six cases, in the talonavicular region in one and the knee in one. Carr⁴ said that he had only one such case, with the ankle and tarsal joints affected, in a series of 100.

Fractures and epiphyseal separations are a common complication, occurring with apparently minimal trauma, in many of these children.^{23,36-40} Cases have been reported by Gillies and Hartung³⁶ in which the callus was so exuberant that it was mistaken for sarcoma on the basis of x-ray visualization and biopsy. These fractures are often virtually painless and the only presenting signs are swelling and heat. In 1965, Freehafer and Mast⁴¹ reported their experience with lower-extremity fractures in patients with spinal cord injury. There were 46 fractures in 23 such patients. Based on this experience, the authors advocated treatment with pillow splints and well-padded plaster casts. For hip fractures they advocated positioning of the hip only, without internal fixation.

Habilitation

With regard to habilitation of these patients, there was for many years unjustified pessimism. This was reinforced by Jaeger's⁴² article in 1953, in which he stated that in studying 68 case histories he was able to find no survivors over the age of 10; he attributed these early deaths to urologic complications. It has become increasingly apparent, however, that with vigorous neurosurgical, urologic and orthopedic management many of these patients can become self-sufficient. As early as 1959, Norton and Foley⁴³ in a study of paraplegia in children found that of 26 patients, 19 of whom were over 18 years of age, 13 were self-supporting. This group included postnatal as well as congenital paraplegics. Independence, once it is achieved by these young paraplegics, appears to be permanent. Bluestone and Deaver,³⁵ Norton and Foley,⁴³ and Sharrard⁴⁴ have all made the point that remarkable degrees of self-sufficiency have been achieved by many of these children. Sharrard contends that more than 80 percent can lead an independent existence, although the average number of orthopedic procedures required in the lower limb was between six and eight in his series. The capacity of paraplegics for sustained ambulation is limited by the high energy expenditure required. Gordon and Vanderwalde,⁴⁵ in a study of this problem, found that the energy ceiling is approximately five to six times the basal metabolic rate; this value approaches the athlete's tolerance for sustained physical activity and is limited in the paraplegic by the decreased absolute capacity for anaerobic work resulting from the small muscle mass of the arms. When the lesion

is at low lumbar and sacral levels, bracing can be minimal and the patient should be completely independent in all activities. Employment is limited only by moderate deficiencies in the activities of ambulation and elevation in such persons. In those with upper lumbar level lesions, only about half will remain functionally ambulatory, and wheelchair independence is a more realistic goal.⁴⁶⁻⁵⁰

Efforts directed toward the goals of a straight spine, straight legs and feet, mobile hips, and, if possible, the development of a pull-to or swing-to gait should be started as soon after birth as possible. Corrective measures for knee, foot, and ankle deformities should not be deferred. Soft-tissue releases may be indicated in certain children under the age of one year. In addition, abduction casting and splinting in an effort to avoid the complication of paralytic dislocation of the hip may be indicated during the first year of life, although this problem remains unresolved at the present. Tendon transfers and neurectomies to correct imbalance are indicated throughout childhood; rotational osteotomy, triple arthrodesis, and spinal fusion may be necessary in some older children.

Upper-extremity strengthening by having the patient walk on his hands while someone holds his legs "wheelbarrow" fashion, and chin-ups should be begun as early as cooperation can be obtained from the child. There are no data of significance as to how early in life these children can be trained in walking. We favor the assumption of the upright posture, either sitting or standing, between six and twelve months of age; fitting for such braces as are indicated; and gait training in parallel bars, proceeding to crutches between one and two years of age. With quadriceps weakness, long leg braces will be required. Results of lesions at the third lumbar level and below, with weakness below the knee only, can be managed with short leg braces if deformity is avoided. Rotational instability of the hip can be a problem in some patients who have adequate quadriceps strength to obviate the need for long leg braces. We have had success with the use of cable twistors attached to the shoes, along with "shoe-horn" laminated plastic inserts for flail feet in some patients, thus avoiding the need for double-upright long leg braces with pelvic band and drop-lock hips. Some loss of stability is conceded in exchange for the much lighter and more easily donned apparatus. Even with lesions at levels as high as the twelfth thoracic segment, we favor such bracing as appears necessary and gait training in early childhood,

recognizing that many of these children will not remain ambulatory as they get older, particularly if they become obese, and that they may find that their high energy expenditures and slow forward progression militate against continued effective ambulation. It is our feeling that bone structure may thus benefit, and perhaps minimize, osteoporosis of disuse and subsequent incidence of fractures. This is another area in which no definitive data have been published.

Summary

The orthopedic and habilitative aspects of the management of patients with spina bifida is complex, involving deformities of the spine, pelvis, hips and lower extremities, and optimal treatment remains undefined in many areas. It is clear, however, that previous pessimism regarding the ultimate self-sufficiency of many of these patients is not justified at present. Early and vigorous orthopedic management can make a definite contribution to the long-term welfare of these unfortunate children.

Addendum

In the time between the presentation of these rounds and their publication, a number of papers and several texts have appeared which have made significant contributions to the orthopedic management and habilitation of myelomeningocele. The American Academy of Orthopaedic Surgeons has published a Symposium on Myelomeningocele,⁵¹ as have several other groups.^{52,53} In addition Menelaus⁵⁴ has written an excellent monograph. Other significant contributions from the orthopedic literature are summarized here.

Sharrard, with a large experience in England,⁵⁵ has recently reviewed his criteria for surgical operation of the hip, knee, foot and spine. He remains enthusiastic about the treatment of children with myelomeningocele, while tempering some of his earlier enthusiasm for immediate closure of spinal defects in all children.

Eighteen patients from a crippled children's hospital (cared for between 1943 and 1959) were followed-up to evaluate functional ability at ages 17 to 31 years.⁵⁶ There were 17 survivors, of whom 12 were termed "competitive" (defined as being independent in activities of daily living and either self-supporting or attending school with good prospect of becoming self-supporting). Four were dependent but not institutionalized and one was confined to a wheelchair in an institution at

age 26 years. Fifteen of the 18 had required surgical procedures to eliminate pressure sores; a total of 107 orthopedic or skin-related surgical procedures had been required among the 18 patients. There was no clear relationship between either the level of paralysis or the intelligence quotient and competitive ability.

The subject of paralytic vertical talus has received increasing attention because it occurs in children with functioning fifth lumbar level nerve roots, a level which permits good ultimate ambulatory capacity. Walker and Cheong-Leen⁵⁷ opened a discussion on its treatment, at the 1973 spring meeting of the British Orthopaedic Association, which is informative to those who must deal with this problem. It indicates a considerable diversity of opinion as to optimal management as well as pathogenesis of this deformity.

Duckworth and Smith⁵⁸ agree with those who believe that imbalance between the evertors and the invertors of the foot is the cause of this deformity, with the peronei acting strongly with the ankle and toe dorsiflexors to overpull the tibialis posterior and the intrinsic foot musculature. They compared the results of release procedures alone with those of combined release and transfer of the peroneus brevis to the tibialis posterior and the tibialis anterior to the neck of the talus, and concluded that there is at present no consistently satisfactory operation when neuromuscular imbalance exists. Two out of three patients were improved, however.

Kopits⁵⁹ has extensively reviewed orthopedic problems and habilitation and suggests decancellation of the talus and cuboid as originally practiced by Verebelyi and Ogston for the management of rigid paralytic clubfoot.

It has been shown by Hay and Walker⁶⁰ that plantar pressures in the feet of children with myelomeningocele are substantially higher than in the feet of age- and sex-matched normal controls. They attribute this in part to the smaller size of the feet in spina bifida, and also postulate that feet in which a previous posterior release has been done are in a condition which they term a "flail deformity," wherein the hindfoot receives excessive loading while the forefoot lies limply and escapes its share of the load.

In the management of paralytic dislocation of the hip, London and Nichols⁶¹ reported superior results from transferring the adductor origin from the pubic tubercle to the ischial tuberosity in combination with a modified Mustard iliopsoas trans-

fer in the third, fourth and fifth lumbar levels in myelodysplasia. They compared the results of this procedure to those obtained following Sharrard's posterolateral iliopsoas transfer and came to the conclusion that reduction of the dislocation and a level pelvis were more easily maintained due to improved muscle balance about the hip if the adductor origin was also transferred.

The indications for iliopsoas transfer were questioned by Carroll,⁶² who feels that this transfer weakens hip flexion but is unlikely to provide abductor or extensor power and should not be done in children with high paraplegia or lumbar kyphosis who are unlikely to walk.

Barden, Meyer and Stelling⁶³ followed 29 survivors of a group originally consisting of 63 severely involved myelodysplastic patients who had been given care from 1928 to 1951 at a Shriners hospital. All had been ambulatory while in the hospital as children, but only two of nine with twelfth thoracic to second lumbar level function were still walking, whereas 19 of 20 with function at the third lumbar level had remained ambulatory. Of considerable interest is the observation that the final position of the hips did not correlate with the ultimate ambulatory status; seven of these 20 patients were ambulatory in spite of persistent hip dislocations. About half of the entire group had scoliosis, and a third of the survivors were self-supporting in full-time employment.

In a small series of young patients, McKibbin⁶⁴ has been able to use a specially designed abduction splint effectively to maintain reduction of paralytic dislocation of the hip. He suggests that this may be a useful adjunct to treatment in the first year of life in those patients with preserved flexor and adductor groups (second and third lumbar levels) until the prognosis for the individual has been assessed.

Sriram, Bobechko and Hall⁶⁵ reviewed data on 33 patients who had been treated for spinal deformities by operation and who had been re-evaluated from a year to 11 years later. Twenty-nine had had posterior spinal fusion, and 26 of these had had Harrington instrumentation as well. Infections and pseudarthroses were common as well as difficult complications to manage, and good results were obtained in only half the patients. They advocate two-stage fusion if the skin is badly scarred or if a residual sac requires excision. This article underscores one of several remaining problem areas in the management of myelomeningocele.

Menelaus⁶⁶ has joined those who advocate osteotomy-resection of kyphosis at the time of posterior closure, which is facilitated by the excision of protruding bone. He states this can be done up to the age of four years, and further states that fusion was obtained in all cases treated in this way.

An improved shortleg brace, made of plastazote-lined polypropylene, for children with third to fifth lumbar level paraplegia has been successfully used by Lindseth and Glancy⁶⁷ in 47 patients. The authors believe that this device brought about considerable improvement in the gaits of nearly all the children and they further state that it represents a major advancement in bracing at this neurological level. Carroll⁶⁸ has recently reviewed current concepts in bracing and the use of carts and standing devices.

REFERENCES

1. Doran PA, Guthkelch AN: Studies in spina bifida cystica—I. General survey and reassessment of the problem. *J Neurol Neurosurg Psychiatry* 24:331-345, Nov 1961
2. Piggot J: Leg movements after closure of myelomeningocele. *J Bone Joint Surg [Br]* 53-B:758, Nov 1971
3. Stark GD, Baker GCW: The neurological involvement of the lower limbs in myelomeningocele. *Dev Med Child Neurol* 9:732-744, Dec 1967
4. Carr TL: The orthopaedic aspects of one hundred cases of spina bifida. *Postgrad Med J* 32:201-210, Apr 1956
5. Hayes JT, Gross HP, Dow S: Surgery for paralytic defects secondary to myelomeningocele and myelodysplasia—An Instructional Course Lecture, The American Academy of Orthopaedic Surgeons. *J Bone Joint Surg [Am]* 46-A:1577-1597, Oct 1964
6. Sharrard WJW, Grosfield I: Management of deformity and paralysis of the foot in myelomeningocele. *J Bone Joint Surg [Br]* 50-B:456-465, Aug 1968
7. Sharrard WJW: The mechanism of paralytic deformity in spina bifida. *Dev Med Child Neurol* 4:310-313, Jun 1962
8. Sharrard WJW: A symposium on paralysis—Part II. Spina bifida. *Physiotherapy* 50:44-49, Feb 1964
9. Sharrard WJW: Paralytic deformity in the lower limb. *J Bone Joint Surg [Br]* 49-B:731-747, Nov 1967
10. Tzimas NA: Orthopedic care of the child with spina bifida. In Swinyard CA (Ed): *Comprehensive Care of the Child with Spina Bifida Manifesta*. New York, Institute of Rehabilitation Medicine, New York University Medical Center, 1966. *Rehabilitation Monograph* 31, pp 45-65
11. Walker G: The early management of varus feet in myelomeningocele. *J Bone Joint Surg [Br]* 53-B:462-467, Aug 1971
12. Menelaus MB: Talcotomy for equinovarus deformity in arthrogryposis and spina bifida. *J Bone Joint Surg [Br]* 53-B:468-473, Aug 1971
13. Drennan JC, Sharrard WJW: The pathological anatomy of convex pes valgus. *J Bone Joint Surg [Br]* 53-B:455-461, Aug 1971
14. Sharrard WJW: Posterior iliopsoas transplantation in the treatment of paralytic dislocation of the hip. *J Bone Joint Surg [Br]* 46-B:426-444, Aug 1964
15. Menelaus MB: Dislocation and deformity of the hip in children with spina bifida cystica. *J Bone Joint Surg [Br]* 51-B:238-251, May 1969
16. Carroll NC, Sharrard WJW: Long-term follow-up of posterior iliopsoas transplantation for paralytic dislocation of the hip. *J Bone Joint Surg [Am]* 54-A:551-560, Apr 1972
17. Salter RB: Panel discussion at the Current Concepts in Children's Orthopaedics Program, American Academy of Orthopaedic Surgeons, Aug 1972
18. Rueda J, Carroll NC: Hip instability in patients with myelomeningocele. *J Bone Joint Surg [Br]* 54-B:422-431, Aug 1972
19. Cruess RL, Turner NS: Paralysis of hip abductor muscles in spina bifida: Results of treatment by the Mustard procedure. *J Bone Joint Surg [Am]* 52-A:1364-1372, Oct 1970
20. Freehafer AA, Vessely JC, Mack RP: Iliopsoas muscle transfer in the treatment of myelomeningocele patients with paralytic hip deformities. *J Bone Joint Surg [Am]* 54-A:1715-1729, Dec 1972
21. Broome HL, Basmajian JV: Survival of iliopsoas muscle after Sharrard procedure: An electromyographic study. *Am J Phys Med* 50:301-302, Dec 1971

MYELOMENINGOCELE

22. Jones GB: Paralytic dislocation of the hip. *J Bone Joint Surg [Br]* 44-B:573-587, Aug 1962
23. Somerville EW: Paralytic dislocation of the hip. *J Bone Joint Surg [Br]* 41-B:279-288, May 1959
24. Donaldson WF: Orthopaedic management of spina bifida. Neuromuscular diseases in children. University of California Continuing Education Course, 1968
25. Hogshead HP, Ponseti IV: Fascia lata transfer to the erector spinae for the treatment of flexion-abduction contractures of the hip in patients with poliomyelitis and meningomyelocele. *J Bone Joint Surg [Am]* 46-A:1389-1404, 1450, Oct 1964
26. McKibbin B: Conservative management of paralytic dislocation of the hip in meningomyelocele. *J Bone Joint Surg [Br]* 53-B:758, Nov 1971
27. Kilfoyle RM, Foley JJ, Norton PL: Spine and pelvic deformity in childhood and adolescent paraplegia: A study of 104 cases. *J Bone Joint Surg [Am]* 47-A:659-682, June 1965
28. Raycroft JF, Curtis BH: Spinal curvatures in myelomeningocele: Natural history and etiology. Paper read at the Thirty-Ninth Annual Meeting of the American Academy of Orthopaedic Surgeons, Feb 1972. Abstract: *J Bone Joint Surg [Am]* 54-A:1335, Sep 1972
29. Sharrard WJW: Spinal osteotomy for congenital kyphosis in myelomeningocele. *J Bone Joint Surg [Br]* 50-B:466-471, Aug 1968
30. Sharrard WJW, Drennan JC: Osteotomy-excision of the spine for lumbar kyphosis in older children with myelomeningocele. *J Bone Joint Surg [Br]* 54-B:50-60, Feb 1972
31. James CCM, Lassman LP: Spinal dysraphism: The diagnosis and treatment of progressive lesions in spina bifida occulta. *J Bone Joint Surg [Br]* 44-B:828-840, Nov 1962
32. Eyring EJ, Wanken JJ: Spinal osteotomy for kyphosis in myelomeningocele. Paper read at the Thirty-Ninth Annual Meeting of the American Academy of Orthopaedic Surgeons, Feb 1972
33. MacEwen GD: Panel discussion at the Current Concepts in Children's Orthopaedics Program, American Academy of Orthopaedic Surgeons, Aug 1972
34. Eckstein HB, Vora RM: Spinal osteotomy for severe kyphosis in children with myelomeningocele. *J Bone Joint Surg [Br]* 54-B:328-333, May 1972
35. Bluestone SS, Deaver GG: Habilitation of the child with spina bifida and myelomeningocele. *JAMA* 161:1248-1251, Jul 28, 1956
36. Gillies CL, Hartung W: Fracture of the tibia in spina bifida vera: Report of two cases. *Radiology* 31:621-623, 1938
37. Golding C: Museum pages—III. Spina bifida and epiphyseal displacement. *J Bone Joint Surg [Br]* 42-B:387-389, May 1960
38. Jeannopoulos CL: Bone changes in children with lesions of the spinal cord or roots. *NY State J Med* 54:3219-3224, Dec 1954
39. Katz JF: Spontaneous fractures in paraplegic children. *J Bone Joint Surg [Am]* 35-A:220-226, Jan 1953
40. Soutter FE: Spina bifida and epiphyseal displacement: Report of two cases. *J Bone Joint Surg [Br]* 44-B:106-109, Feb 1962
41. Freehafer AA, Mast WA: Lower extremity fractures in patients with spinal-cord injury. *J Bone Joint Surg [Am]* 47-A:683-694, Jun 1965
42. Jaeger R: Congenital spinal meningocele. *JAMA* 153:792-795, Oct 31, 1953
43. Norton PL, Foley JJ: Paraplegia in children. *J Bone Joint Surg [Am]* 41-A:1291-1309, Oct 1959
44. Sharrard WJW: Rehabilitation problems in spina bifida. *Bull Schweiz Akad Med Wiss* 28:61-74, Apr 1972
45. Gordon EE, Vanderwalde H: Energy requirements in paraplegic ambulation. *Arch Phys Med Rehabil* 37:276-285, May 1956
46. Arthur AB, Bush RT, Guard F, et al: Spina bifida cystica—Survey of 107 cases. *NZ Med J* 75:272-277, May 1972
47. Lorber J: Results of treatment of myelomeningocele—An analysis of 524 unselected cases, with special reference to possible selection for treatment. *Dev Med Child Neurol* 13:279-303, Jun 1971
48. Richings JC, Eckstein HB: Locomotor and educational achievements of children with myelomeningocele. *Ann Phys Med* 10:291-298, May 1970
49. Tzimas NA, Badell-Ribera A: Orthopedic and habilitation management of patients with spina bifida and myelomeningocele. *Med Clin North Am* 53:502-509, May 1969
50. Hoffer MM, Feiwell E, Perry R, et al: Functional ambulation in patients with myelomeningocele. *J Bone Joint Surg [Am]* 55-A:137-148, Jan 1973
51. American Academy of Orthopaedic Surgeons: Symposium on Myelomeningocele. St. Louis, Mo, C V Mosby Company, 1972
52. Freeman JM (Ed): Practical Management of Meningomyelocele. Baltimore, University Press, 1974
53. Bunch WH, Cass AS, Bensman AS, Long DM: Modern Management of Myelomeningocele. St. Louis, Mo, Warren H Green Inc, 1972, pp 121-209
54. Menelaus MB: The Orthopaedic Management of Spina Bifida Cystica. Edinburgh, Livingstone, 1971
55. Sharrard WJW: The orthopaedic surgery of spina bifida. *Clin Orthop* 92:195-213, 1973
56. Levin GD: Functional evaluation of eighteen adult myelomeningocele patients. *Clin Orthop* 100:101-107, 1974
57. Walker GF, Cheong-Leen P: The surgical management of paralytic vertical talus in myelomeningocele. *J Bone Joint Surg [Br]* 55B:876-877, Nov 1973
58. Duckworth T, Smith TWD: The treatment of paralytic convex pes valgus. *J Bone Joint Surg [Br]* 56B:305-313, May 1974
59. Kopits SE: Orthopedic aspects of meningomyelocele. In: Practical Management of Meningomyelocele. Freeman JM (Ed), Baltimore, University Press, 1974
60. Hay MC, Walker G: Plantar pressures in healthy children and in children with myelomeningocele. *J Bone Joint Surg [Br]* 55:828-833, Nov 1973
61. London JT, Nichols O: Paper read at the Forty-First Annual Meeting of the American Academy of Orthopaedic Surgeons, Jan 1974
62. Carroll NC: Hip instability in patients with myelomeningocele. *J Bone Joint Surg [Br]* 55B:661, Aug 1973
63. Barden GA, Meyer LC, Stelling FH: Paper read at the Forty-First Annual Meeting of the American Academy of Orthopaedic Surgeons, Jan 1974
64. McKibbin B: The use of splintage in the management of paralytic dislocation of the hip in spina bifida cystica. *J Bone Joint Surg [Br]* 55B:163-172, Feb 1973
65. Sriram K, Bobechko WP, Hall JE: Surgical management of spinal deformities in spina bifida. *J Bone Joint Surg [Br]* 54B:666-676, Nov 1972
66. Menelaus M: Spinal deformity in spina bifida. *J Bone Joint Surg [Br]* 55B:223-224, Feb 1973
67. Lindseth RE, Glancy J: Polypropylene lower-extremity braces for paraplegia due to myelomeningocele. *J Bone Joint Surg [Am]* 56A:556-563, Apr 1974
68. Carroll N: The orthotic management of the spina bifida child. *Clin Orthop* 102:108-114, 1974

Refer to: Goodner EK: Myelomeningocele—Part II: Ophthalmologic problems. In: Myelomeningocele—A symposium. *West J Med* 121:291-292, Oct 1974

MYELOMENINGOCELE—PART II

Ophthalmologic Problems

ERNEST K. GOODNER, MD, *San Francisco*

DEVELOPMENTAL abnormalities of the eye may occur in association with defects in the formation of the midline portions of the cranium and spinal column. These ocular anomalies are severe, rare and generally not susceptible to treatment or repair. Fortunately, the majority of children with spina bifida and related conditions have normal eyes and visual development.

The ocular abnormalities that occur in association with malformations of the cranium and vertebral column are produced generally in one of two ways: (1) Defects in ocular function may result from the direct effect of maldevelopment in the cranium or vertebrae on underlying neural components of the extraocular visual system (for ex-

Part II of an article in six parts.

From the Department of Ophthalmology, University of California, San Francisco.

Reprint requests to: E. K. Goodner, MD, Department of Ophthalmology, University of California, San Francisco, San Francisco, CA 94143.